Hemifacial Spasm

Caetano Coimbra, MD; Grant Gilliland, MD; Manu Gupta, MD; Amol Bhatki, MD; Yoav Hahn, MD

Hemifacial spasm (HFS) is a condition of unilateral, involuntary, irregular spasmodic movements of the face. The condition is most commonly a result of vascular loop compression at the root entry zone of the facial nerve. HFS occurs in approximately 11 per 100,000 individuals and is more common in middle age, affecting women more often than men.

HFS symptoms generally start at the periorbital muscles with intermittent muscle activity under and around the eye and progresses to involve the muscles at the midface and perioral area. The condition tends to evolve slowly and patients often come to medical attention many years after the onset of the symptoms. The facial spasm is generally more pronounced during stressful situations and can cause emotional and physical distress to the patient. Over time, the affected individual tends to become self conscious of their condition which often results in insecurity and social withdrawal, interfering with daily activities and professional life. When HFS is pronounced it can cause difficulty focusing with the ipsilateral eye, which can affect reading and driving. Persistent spasm can result in droopiness of the ipsilateral eyelid due to dehiscence or stretching of the levator aponeurosis. The spasm can cause an induced astigmatism of the cornea with resultant blurred vision. In addition, because of the hypertrophic sphincter muscle of the eye, abnormal tearing can result.

HFS is most commonly caused by vascular compression of the facial nerve (FN) at the root entry zone (REZ) near the brainstem (fig 1). MRI utilizing high resolution facial nerve protocol allows precise visualization of the offending artery and its relation to the FN. Less often, HFS can be the result of FN compression by tumors occupying the cerebello-pontine angle, including an epidermoid (fig 2). FN injury, including trauma to the nerve or viral infection causing facial paralysis, can also cause HFS. In these cases the facial spasm tend to start after partial recovery of the facial paralysis.

The Skull Base Center at BUMC uses a multidisciplinary team of professionals on its medical staff including a Neurosurgeon, Oculoplastic surgeon, Neurootologist and Neuroradiologist for comprehensive evaluation and customized approach for patients with HFS. The typical work up for these patients includes audiogram, FN electromyogram, FN protocol MRI, and consultation with our multidisciplinary team.

Treatment for patients with HFS may include palliative botox injection or microvascular decompression (MVD). Botulinum neurotoxin A is an effective neuromuscular blocking agent.

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It acts by blocking the nerve signal to the affected muscle resulting in relaxation of the muscle. It has an effectiveness rate of approximately 95% with few side effects. Side effects may include droopy eyelid, retraction of the lower eyelid, abnormal tearing, double vision, poor exercise tolerance and allergic reaction. Botulinum neurotoxin A typically lasts 2-4 months at which time injections will need to be repeated.

Surgical treatment for HFS includes a modified focused retro-sigmoid occipital approach. This modified approach has eliminated the post-op chronic unilateral headache associated with the traditional sub-occipital approach. This modified approach allows access under the cerebellum to the IX, X, and XI lower cranial nerves (LCN) at the vicinity of the Foramen Luschka of the 4th ventricle. (Figure 3) Next, the choroidal plexus is identified and more superiorly the origin of the acoustic nerve (AN) is visualized at the brainstem. The vascular compression of the nerve at the REZ is medial to the AN. The vascular compression can be caused by the AICA, the PICA or the vertebral artery. The offending vessel is mobilized away from the FN and a sponge is placed between the nerve and the artery. (Figure 4) We have found that this more inferior approach allows direct access to the REZ of the AN and FN and therefore avoids manipulation of the nerves in the cerebello-pontine angle thus preventing complications of post-operative facial paralysis and/or hearing loss.

Intraoperative monitoring of the AN and FN is critical to successful surgery. FN lateral spread potentials are measured intraoperatively and disappearance of this lateral spread response can be used as a predictor of decompression of the nerve. After successful decompression, intraoperative lateral spread potential is eliminated predicting resolution of the HFS.

In summary, HFS is a debilitating condition that needs to be properly evaluated and treated with a sub specialized multidisciplinary team. Botox injection is a viable, less invasive treatment option which can provide temporary relief. When surgery is needed, MVD of the FN is an effective way of obtaining a more permanent solution to HFS. Knowledge of the complex anatomy of this region in the skull base is paramount to obtain successful outcomes and avoid potential complications of the procedure. Customized modifications of surgical techniques developed at the BUMC Skull Base Center have allowed favorable sustained outcomes for patients with HFS.